

#2

OIPE

## RAW SEQUENCE LISTING

DATE: 07/23/2001

PATENT APPLICATION: US/09/895,072

TIME: 13:19:44

Input Set : A:\210119US0CONT.txt

Output Set: N:\CRF3\07232001\I895072.raw

3 &lt;110&gt; APPLICANT: CANFIELD, WILLIAM M

5 &lt;120&gt; TITLE OF INVENTION: METHODS FOR PRODUCING HIGHLY PHOSPHORYLATED LYSOSOMAL

## HYDROLASES

7 &lt;130&gt; FILE REFERENCE: 210119US0CONT

C--&gt; 9 &lt;140&gt; CURRENT APPLICATION NUMBER: US/09/895,072

C--&gt; 9 &lt;141&gt; CURRENT FILING DATE: 2001-07-02

9 &lt;150&gt; PRIOR APPLICATION NUMBER: 60/153,831

10 &lt;151&gt; PRIOR FILING DATE: 1999-09-14

12 &lt;150&gt; PRIOR APPLICATION NUMBER: US 09/635,872

13 &lt;151&gt; PRIOR FILING DATE: 2000-08-10

15 &lt;160&gt; NUMBER OF SEQ ID NOS: 52

17 &lt;170&gt; SOFTWARE: PatentIn version 3.1

19 &lt;210&gt; SEQ ID NO: 1

20 &lt;211&gt; LENGTH: 928

21 &lt;212&gt; TYPE: PRT

22 &lt;213&gt; ORGANISM: Homo sapiens

24 &lt;400&gt; SEQUENCE: 1

26 Met Leu Phe Lys Leu Leu Gln Arg Gln Thr Tyr Thr Cys Leu Ser His  
 27 1 5 10 15  
 30 Arg Tyr Gly Leu Tyr Val Cys Phe Leu Gly Val Val Val Thr Ile Val  
 31 20 25 30  
 34 Ser Ala Phe Gln Phe Gly Glu Val Val Leu Glu Trp Ser Arg Asp Gln  
 35 35 40 45  
 38 Tyr His Val Leu Phe Asp Ser Tyr Arg Asp Asn Ile Ala Gly Lys Ser  
 39 50 55 60  
 42 Phe Gln Asn Arg Leu Cys Leu Pro Met Pro Ile Asp Val Val Tyr Thr  
 43 65 70 75 80  
 46 Trp Val Asn Gly Thr Asp Leu Glu Leu Leu Lys Glu Leu Gln Gln Val  
 47 85 90 95  
 50 Arg Glu Gln Met Glu Glu Glu Gln Lys Ala Met Arg Glu Ile Leu Gly  
 51 100 105 110  
 54 Lys Asn Thr Thr Glu Pro Thr Lys Lys Ser Glu Lys Gln Leu Glu Cys  
 55 115 120 125  
 58 Leu Leu Thr His Cys Ile Lys Val Pro Met Leu Val Leu Asp Pro Ala  
 59 130 135 140  
 62 Leu Pro Ala Asn Ile Thr Leu Lys Asp Val Pro Ser Leu Tyr Pro Ser  
 63 145 150 155 160  
 66 Phe His Ser Ala Ser Asp Ile Phe Asn Val Ala Lys Pro Lys Asn Pro  
 67 165 170 175  
 70 Ser Thr Asn Val Ser Val Val Val Phe Asp Ser Thr Lys Asp Val Glu  
 71 180 185 190  
 74 Asp Ala His Ser Gly Leu Leu Lys Gly Asn Ser Arg Gln Thr Val Trp  
 75 195 200 205  
 78 Arg Gly Tyr Leu Thr Thr Asp Lys Glu Val Pro Gly Leu Val Leu Met  
 79 210 215 220  
 82 Gln Asp Leu Ala Phe Leu Ser Gly Phe Pro Pro Thr Phe Lys Glu Thr  
 83 225 230 235 240  
 86 Asn Gln Leu Lys Thr Lys Leu Pro Glu Asn Leu Ser Ser Lys Val Lys

ENTERED

See page 5

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87          245          250          255
90 Leu Leu Gln Leu Tyr Ser Glu Ala Ser Val Ala Leu Leu Lys Leu Asn
91          260          265          270
94 Asn Pro Lys Asp Phe Gln Glu Leu Asn Lys Gln Thr Lys Lys Asn Met
95          275          280          285
98 Thr Ile Asp Gly Lys Glu Leu Thr Ile Ser Pro Ala Tyr Leu Leu Trp
99          290          295          300
102 Asp Leu Ser Ala Ile Ser Gln Ser Lys Gln Asp Glu Asp Ile Ser Ala
103 305          310          315          320
106 Ser Arg Phe Glu Asp Asn Glu Glu Leu Arg Tyr Ser Leu Arg Ser Ile
107          325          330          335
110 Glu Arg His Ala Pro Trp Val Arg Asn Ile Phe Ile Val Thr Asn Gly
111          340          345          350
114 Gln Ile Pro Ser Trp Leu Asn Leu Asp Asn Pro Arg Val Thr Ile Val
115          355          360          365
118 Thr His Gln Asp Val Phe Arg Asn Leu Ser His Leu Pro Thr Phe Ser
119          370          375          380
122 Ser Pro Ala Ile Glu Ser His Ile His Arg Ile Glu Gly Leu Ser Gln
123 385          390          395          400
126 Lys Phe Ile Tyr Leu Asn Asp Asp Val Met Phe Gly Lys Asp Val Trp
127          405          410          415
130 Pro Asp Asp Phe Tyr Ser His Ser Lys Gly Gln Lys Val Tyr Leu Thr
131          420          425          430
134 Trp Pro Val Pro Asn Cys Ala Glu Gly Cys Pro Gly Ser Trp Ile Lys
135          435          440          445
138 Asp Gly Tyr Cys Asp Lys Ala Cys Asn Asn Ser Ala Cys Asp Trp Asp
139          450          455          460
142 Gly Gly Asp Cys Ser Gly Asn Ser Gly Gly Ser Arg Tyr Ile Ala Gly
143 465          470          475          480
146 Gly Gly Gly Thr Gly Ser Ile Gly Val Gly His Pro Trp Gln Phe Gly
147          485          490          495
150 Gly Gly Ile Asn Ser Val Ser Tyr Cys Asn Gln Gly Cys Ala Asn Ser
151          500          505          510
154 Trp Leu Ala Asp Lys Phe Cys Asp Gln Ala Cys Asn Val Leu Ser Cys
155          515          520          525
158 Gly Phe Asp Ala Gly Asp Cys Gly Gln Asp His Phe His Glu Leu Tyr
159          530          535          540
162 Lys Val Ile Leu Leu Pro Asn Gln Thr His Tyr Ile Ile Pro Lys Gly
163 545          550          555          560
166 Glu Cys Leu Pro Tyr Phe Ser Phe Ala Glu Val Ala Lys Arg Gly Val
167          565          570          575
170 Glu Gly Ala Tyr Ser Asp Asn Pro Ile Ile Arg His Ala Ser Ile Ala
171          580          585          590
174 Asn Lys Trp Lys Thr Ile His Leu Ile Met His Ser Gly Met Asn Ala
175          595          600          605
178 Thr Thr Ile His Phe Asn Leu Thr Phe Gln Asn Thr Asn Asp Glu Glu
179          610          615          620
182 Phe Lys Met Gln Ile Thr Val Glu Val Asp Thr Arg Glu Gly Pro Lys
183 625          630          635          640

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```

186 Leu Asn Ser Thr Ala Gln Lys Gly Tyr Glu Asn Leu Val Ser Pro Ile
187           645           650           655
190 Thr Leu Leu Pro Glu Ala Glu Ile Leu Phe Glu Asp Ile Pro Lys Glu
191           660           665           670
194 Lys Arg Phe Pro Lys Phe Lys Arg His Asp Val Asn Ser Thr Arg Arg
195           675           680           685
198 Ala Gln Glu Glu Val Lys Ile Pro Leu Val Asn Ile Ser Leu Leu Pro
199           690           695           700
202 Lys Asp Ala Gln Leu Ser Leu Asn Thr Leu Asp Leu Gln Leu Glu His
203 705           710           715           720
206 Gly Asp Ile Thr Leu Lys Gly Tyr Asn Leu Ser Lys Ser Ala Leu Leu
207           725           730           735
210 Arg Ser Phe Leu Met Asn Ser Gln His Ala Lys Ile Lys Asn Gln Ala
211           740           745           750
214 Ile Ile Thr Asp Glu Thr Asn Asp Ser Leu Val Ala Pro Gln Glu Lys
215           755           760           765
218 Gln Val His Lys Ser Ile Leu Pro Asn Ser Leu Gly Val Ser Glu Arg
219           770           775           780
222 Leu Gln Arg Leu Thr Phe Pro Ala Val Ser Val Lys Val Asn Gly His
223 785           790           795           800
226 Asp Gln Gly Gln Asn Pro Pro Leu Asp Leu Glu Thr Thr Ala Arg Phe
227           805           810           815
230 Arg Val Glu Thr His Thr Gln Lys Thr Ile Gly Gly Asn Val Thr Lys
231           820           825           830
234 Glu Lys Pro Pro Ser Leu Ile Val Pro Leu Glu Ser Gln Met Thr Lys
235           835           840           845
238 Glu Lys Lys Ile Thr Gly Lys Glu Lys Glu Asn Ser Arg Met Glu Glu
239           850           855           860
242 Asn Ala Glu Asn His Ile Gly Val Thr Glu Val Leu Leu Gly Arg Lys
243 865           870           875           880
246 Leu Gln His Tyr Thr Asp Ser Tyr Leu Gly Phe Leu Pro Trp Glu Lys
247           885           890           895
250 Lys Lys Tyr Phe Gln Asp Leu Leu Asp Glu Glu Glu Ser Leu Lys Thr
251           900           905           910
254 Gln Leu Ala Tyr Phe Thr Asp Ser Lys Asn Thr Gly Arg Gln Leu Lys
255           915           920           925
258 <210> SEQ ID NO: 2
259 <211> LENGTH: 328
260 <212> TYPE: PRT
261 <213> ORGANISM: Homo sapiens
263 <400> SEQUENCE: 2
265 Asp Thr Phe Ala Asp Ser Leu Arg Tyr Val Asn Lys Ile Leu Asn Ser
266 1           5           10           15
269 Lys Phe Gly Phe Thr Ser Arg Lys Val Pro Ala His Met Pro His Met
270           20           25           30
273 Ile Asp Arg Ile Val Met Gln Glu Leu Gln Asp Met Phe Pro Glu Glu
274           35           40           45
277 Phe Asp Lys Thr Ser Phe His Lys Val Arg His Ser Glu Asp Met Gln
278           50           55           60

```

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Output Set: N:\CRF3\07232001\I895072.raw

```

281 Phe Ala Phe Ser Tyr Phe Tyr Tyr Leu Met Ser Ala Val Gln Pro Leu
282 65              70              75              80
285 Asn Ile Ser Gln Val Phe Asp Glu Val Asp Thr Asp Gln Ser Gly Val
286              85              90              95
289 Leu Ser Asp Arg Glu Ile Arg Thr Leu Ala Thr Arg Ile His Glu Leu
290              100             105             110
293 Pro Leu Ser Leu Gln Asp Leu Thr Gly Leu Glu His Met Leu Ile Asn
294              115             120             125
297 Cys Ser Lys Met Leu Pro Ala Asp Ile Thr Gln Leu Asn Asn Ile Pro
298              130             135             140
301 Pro Thr Gln Glu Ser Tyr Tyr Asp Pro Asn Leu Pro Pro Val Thr Lys
302 145              150              155              160
305 Ser Leu Val Thr Asn Cys Lys Pro Val Thr Asp Lys Ile His Lys Ala
306              165              170              175
309 Tyr Lys Asp Lys Asn Lys Tyr Arg Phe Glu Ile Met Gly Glu Glu Glu
310              180             185             190
313 Ile Ala Phe Lys Met Ile Arg Thr Asn Val Ser His Val Val Gly Gln
314              195             200             205
317 Leu Asp Asp Ile Arg Lys Asn Pro Arg Lys Phe Val Cys Leu Asn Asp
318              210             215             220
321 Asn Ile Asp His Asn His Lys Asp Ala Gln Thr Val Lys Ala Val Leu
322 225              230             235             240
325 Arg Asp Phe Tyr Glu Ser Met Phe Pro Ile Pro Ser Gln Phe Glu Leu
326              245             250             255
329 Pro Arg Glu Tyr Arg Asn Arg Phe Leu His Met His Glu Leu Gln Glu
330              260             265             270
333 Trp Arg Ala Tyr Arg Asp Lys Leu Lys Phe Trp Thr His Cys Val Leu
334              275             280             285
337 Ala Thr Leu Ile Met Phe Thr Ile Phe Ser Phe Phe Ala Glu Gln Leu
338              290             295             300
341 Ile Ala Leu Lys Arg Lys Ile Phe Pro Arg Arg Arg Ile His Lys Glu
342 305              310             315             320
345 Ala Ser Pro Asn Arg Ile Arg Val
346              325
349 <210> SEQ ID NO: 3
350 <211> LENGTH: 305
351 <212> TYPE: PRT
352 <213> ORGANISM: Homo sapiens
354 <220> FEATURE:
355 <221> NAME/KEY: SIGNAL
356 <222> LOCATION: (1)..(24)
357 <223> OTHER INFORMATION:
360 <400> SEQUENCE: 3
362 Met Ala Ala Gly Leu Ala Arg Leu Leu Leu Leu Gly Leu Ser Ala
363 1              5              10              15
366 Gly Gly Pro Ala Pro Ala Gly Ala Ala Lys Met Lys Val Val Glu Glu
367              20              25              30
370 Pro Asn Ala Phe Gly Val Asn Asn Pro Phe Leu Pro Gln Ala Ser Arg
371              35              40              45

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DATE: 07/23/2001

TIME: 13:19:44

Input Set : A:\210119US0CONT.txt

Output Set: N:\CRF3\07232001\I895072.raw

```

374 Leu Gln Ala Lys Arg Asp Pro Ser Pro Val Ser Gly Pro Val His Leu
375      50                      55                      60
378 Phe Arg Leu Ser Gly Lys Cys Phe Ser Leu Val Glu Ser Thr Tyr Lys
379 65                      70                      75                      80
382 Tyr Glu Phe Cys Pro Phe His Asn Val Thr Gln His Glu Gln Thr Phe
383                      85                      90                      95
386 Arg Trp Asn Ala Tyr Ser Gly Ile Leu Gly Ile Trp His Glu Trp Glu
387                      100                     105                     110
390 Ile Ala Asn Asn Thr Phe Thr Gly Met Trp Met Arg Asp Gly Asp Ala
391                      115                     120                     125
394 Cys Arg Ser Arg Ser Arg Gln Ser Lys Val Glu Leu Ala Cys Gly Lys
395                      130                     135                     140
398 Ser Asn Arg Leu Ala His Val Ser Glu Pro Ser Thr Cys Val Tyr Ala
399 145                      150                     155                     160
402 Leu Thr Phe Glu Thr Pro Leu Val Cys His Pro His Ala Leu Leu Val
403                      165                     170                     175
406 Tyr Pro Thr Leu Pro Glu Ala Leu Gln Arg Gln Trp Asp Gln Val Glu
407                      180                     185                     190
410 Gln Asp Leu Ala Asp Glu Leu Ile Thr Pro Gln Gly His Glu Lys Leu
411                      195                     200                     205
414 Leu Arg Thr Leu Phe Glu Asp Ala Gly Tyr Leu Lys Thr Pro Glu Glu
415                      210                     215                     220
418 Asn Glu Pro Thr Gln Leu Glu Gly Gly Pro Asp Ser Leu Gly Phe Glu
419 225                      230                     235                     240
422 Thr Leu Glu Asn Cys Arg Lys Ala His Lys Glu Leu Ser Lys Glu Ile
423                      245                     250                     255
426 Lys Arg Leu Lys Gly Leu Leu Thr Gln His Gly Ile Pro Tyr Thr Arg
427                      260                     265                     270
430 Pro Thr Glu Thr Ser Asn Leu Glu His Leu Gly His Glu Thr Pro Arg
431                      275                     280                     285
434 Ala Lys Ser Pro Glu Gln Leu Arg Gly Asp Pro Gly Leu Arg Gly Ser
435                      290                     295                     300
438 Leu
439 305
442 <210> SEQ ID NO: 4
443 <211> LENGTH: 5597
444 <212> TYPE: DNA
445 <213> ORGANISM: Homo sapiens
447 <400> SEQUENCE: 4
448 cggagccgag cgggcgtccg tcgccggagc tgcaatgagc ggcgcgccgga ggctgtgacc      60
450 tgcgcgcggc ggcccgaccg gggcccctga atggcggtc gctgaggcgg cggcggcggc      120
452 ggcggctcag gtcctcggg gcgtggcgtg gcggtgaagg ggtgatgctg ttcaagctcc      180
454 tgcagagaca aacctatacc tgcctgtccc acaggtatgg gctctacgtg tgcttcttgg      240
456 ggcgtcgttg caccatcgtc tccgccttcc agttcggaga ggtggttctg gaatggagcc      300
458 gagatcaata ccatgttttg tttgattcct atagagacaa tattgctgga aagtcctttc      360
460 agaatcggct ttgtctgcc atgccgattg acgttggtta cacctgggtg aatggcacag      420
462 atcttgaact actgaaggaa ctacagcagg tcagagaaca gatggaggag gagcagaaag      480
464 caatgagaga aatccttggg aaaaacacaa cggaacctac taagaagagt gagaagcagt      540
466 tagagtgttt gctaacacac tgcattaagg tgccaatgct tgtactggac ccagccctgc      600

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Use of n and / or Xaa has been detected in the  
 Sequence Listing. Review the Sequence Listing  
 to ensure a corresponding explanation is present  
 in the <220> to <223> fields of each sequence  
 using n or Xaa.

## VERIFICATION SUMMARY

PATENT APPLICATION: US/09/895,072

DATE: 07/23/2001

TIME: 13:19:45

Input Set : A:\210119US0CONT.txt

Output Set: N:\CRF3\07232001\I895072.raw

L:9 M:270 C: Current Application Number differs, Replaced Current Application No

L:9 M:271 C: Current Filing Date differs, Replaced Current Filing Date

L:1120 M:341 W: (46) "n" or "Xaa" used, for SEQ ID#:10

L:2207 M:341 W: (46) "n" or "Xaa" used, for SEQ ID#:17

L:2211 M:341 W: (46) "n" or "Xaa" used, for SEQ ID#:17

L:2213 M:341 W: (46) "n" or "Xaa" used, for SEQ ID#:17

L:2754 M:341 W: (46) "n" or "Xaa" used, for SEQ ID#:23

L:2926 M:341 W: (46) "n" or "Xaa" used, for SEQ ID#:35